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J S Lewin and P Lavertu

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Aggressive Fibromatosis of the Prevertebral and Retropharyngeal Spaces: MR and CT Characteristics

Jonathan S. Lewin and Pierre Lavertu

Summary: A 47-year-old man had aggressive fibromatosis, and CT and MR showed a large, multilobulated soft-tissue mass within the prevertebral and retropharyngeal spaces. On CT, the lesion was slightly higher in attenuation than adjacent muscle; on MR, it was intermediate between muscle and fat on unenhanced T1-weighted images, isointense with fat on intermediate-weighted images, hyperintense relative to fat on T2-weighted images, and markedly enhanced after administration of gadopentetate dimeglumine. Multiple small focal and linear areas of decreased signal intensity that did not enhance with gadopentetate dimeglumine were observed on all pulse sequences.

Index term: Neck, neoplasms

Aggressive fibromatosis (or extraabdominal desmoid) of the neck is a rare nonencapsulated, nonmetastasizing fibroblastic proliferative lesion that is histologically benign but has a high rate of recurrence.

Case Report

A 47-year-old black man was referred to the otolaryngology department for evaluation of a large neck mass. The patient initially was seen more than 3 years previously because of neck stiffness and right shoulder pain. A slight limitation of neck rotation was noted on physical examination and the patient was treated with physical therapy. The discomfort became progressively worse and was most noticeable at night and during exercise. Five months before presentation to our institution, dysphagia developed that progressed until swallowing was possible only with full neck extension. Diffuse firm enlargement of the neck was noted. The patient stated that his shirt collar size had increased 2 inches over the prior 2 years. A cervical spine radiograph showed a significant increase in the prevertebral soft tissues and a reversal of the normal cervical lordosis. A barium esophagogram showed anterior displacement of the esophagus.

The patient underwent contrast-enhanced computed tomography (CT) and magnetic resonance (MR) imaging. MR was performed with a standard clinical 1.5-T imager with T1-weighted, intermediate-weighted, T2-weighted, and gadopentetate dimeglumine–enhanced T1-weighted spin-echo pulse sequences. Both imaging techniques showed a large, multilobulated soft-tissue mass within the prevertebral and retropharyngeal spaces extending from the C-2 vertebral level to the thoracic inlet. It was inseparable from the prevertebral musculature and displaced and compressed the trachea and esophagus anteriorly and carotid sheaths laterally, without vascular encasement (Fig 1). The right longus capitis and longus colli muscles appeared to be replaced by the mass, and although the left prevertebral muscles were visible, the left prevertebral fascial plane was indistinct. On CT, the lesion was slightly higher in attenuation than adjacent muscle, with several mildly heterogeneous areas within. There was no evidence of calcification, bone erosion, cyst formation, necrosis, or associated adenopathy.

Signal intensity on MR was intermediate between muscle and fat on unenhanced T1-weighted images, isointense with fat on intermediate-weighted images, and hyperintense relative to fat on T2-weighted images. Marked enhancement was noted after administration of gadopentetate dimeglumine. The lesion was slightly more heterogenous than on the CT study, with multiple small focal and linear areas of decreased signal intensity on all pulse sequences that did not enhance with gadopentetate dimeglumine.

After the imaging evaluation, the patient underwent transcervical, transsternal excision of the lesion. The mass appeared to arise within the prevertebral muscles on the right, with gross involvement of the prevertebral musculature, fascia, and ligaments and with anterior extension into the retropharyngeal space. The tumor could be separated from neurovascular structures and the visceral content of the neck. An adequate margin could not be obtained surgically, and the patient was discharged home on oral tamoxifen to reduce the likelihood of tumor recurrence.

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From the Department of Radiology, University Hospitals of Cleveland, Ohio (J.S.L.), and Department of Otolaryngology, Cleveland Clinic Foundation (P.L.).

Address reprint requests to Jonathan S. Lewin, MD, Department of Radiology, University Hospitals of Cleveland, 2074 Abington Rd, Cleveland, OH 44106.

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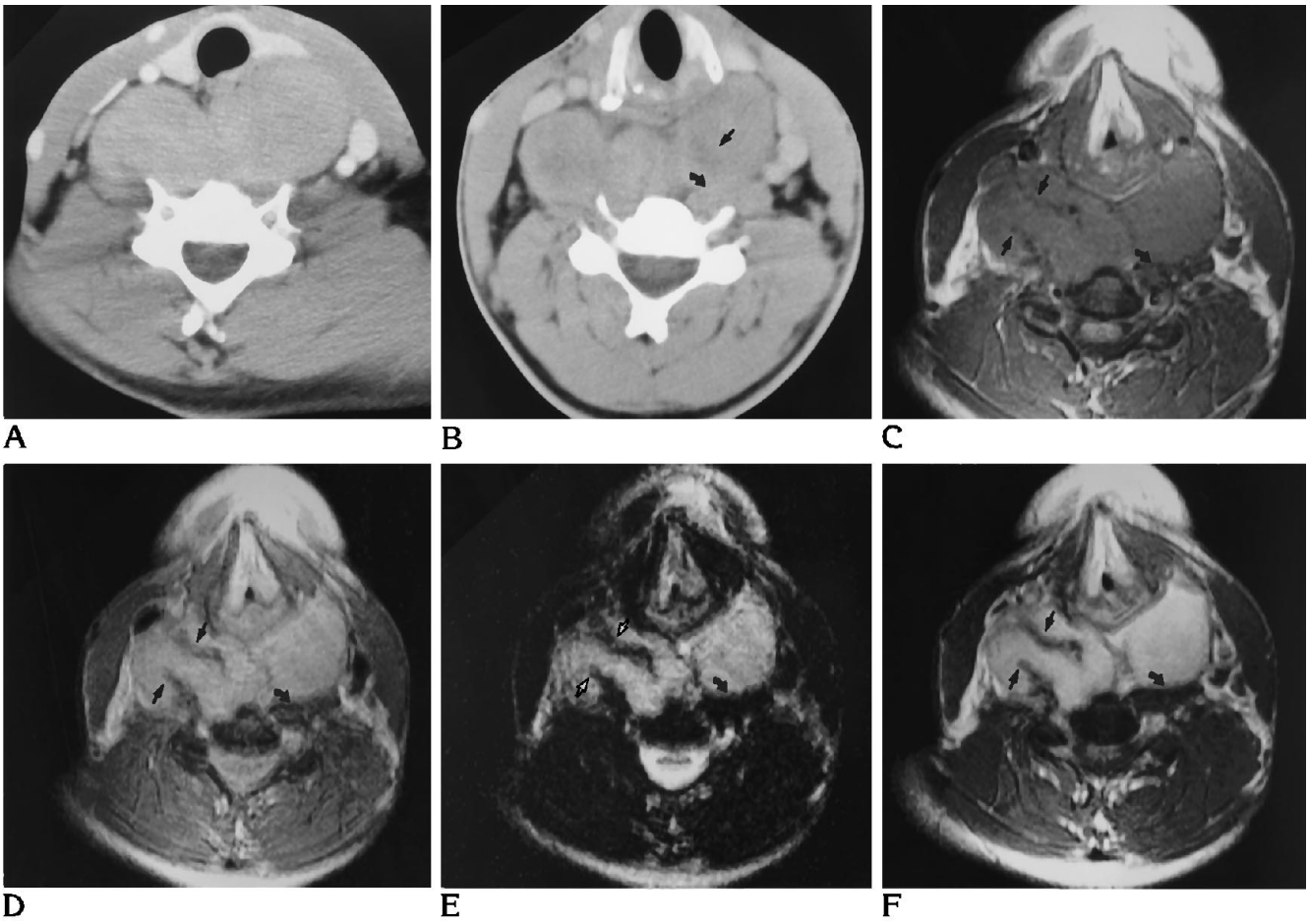

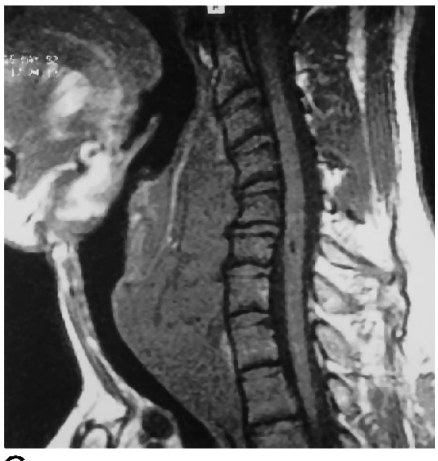

Fig 1. *A* and *B*, Axial contrast-enhanced CT scans show a large mass in the prevertebral/retropharyngeal region. The left prevertebral muscle (*curved arrow*) is identified and appears to be separate from the mass. The right prevertebral muscle cannot be identified and is probably replaced by tumor. Structures in the visceral space are displaced anteriorly and the carotid sheath structures laterally by the lesion. Slight heterogeneity is noted (*arrow*).

C–F, Axial T1-weighted (750/15 [repetition time/echo time/excitations], *C*), intermediate-weighted (3000/22, *D*), T2-weighted (3000/90, *E*), and gadopentetate dimeglumine–enhanced T1-weighted (850/22, *F*) MR images at the level of the thyroid cartilage show similar displacement of anterior and lateral structures. The distinction between mass and compressed left prevertebral muscles (*curved arrows*), as well as replacement of the right prevertebral muscles, is visualized better on MR than on CT. Linear areas of decreased signal intensity noted within the mass on the right (*straight arrows*) on all pulse sequences likely represent areas of fibrosis.

G, Sagittal T1-weighted (425/15) MR image shows the superior-inferior extent of the lesion, kyphosis of the cervical spine, and anterior displacement of the visceral space structures.

Gross pathologic analysis demonstrated multiple, irregularly shaped rubbery white nodules, with small amounts of apparent skeletal muscle attached to several of the larger fragments. Histologically, the lesion was characterized by a proliferation of spindle-shaped cells separated by variable amounts of collagen. The nuclei were relatively uniform and spindle shaped, with inconspicuous nucleoli and with only rare mitotic figures.

Discussion

Aggressive fibromatosis is a soft-tissue neoplasm arising from fascial or musculoaponeurotic structures. It has also been termed *extra-abdominal desmoid*, *musculoaponeurotic fibromatosis*, or *desmoma* (1). Although the tumor was described in 1832, it was Sanger in

1864 who defined the characteristics that distinguish this tumor, including fibroblastic origin, local invasion of muscle and fascia, and lack of a capsule (1). Aggressive fibromatosis is rare, reported to occur in between two and four cases per million inhabitants per year, representing 0.03% to 0.1% of all tumors (1). The age range of affected persons spans from newborns to the elderly, with most cases appearing in the third and fourth decades (2). Several series suggest this process is more common in female subjects (3, 4), whereas others suggest an equal sex distribution (5). The location of aggressive fibromatosis varies from series to series, with an average of approximately 58% in extraabdominal locations and the remainder either within the abdomen or abdominal wall (1). Aggressive fibromatosis in the head and neck has been reported to constitute approximately 11% of extraabdominal cases (3). Although its cause is unknown, several factors such as pregnancy, estrogenic hormones, heredity, and accidental or surgical trauma have been implicated (1).

Aggressive fibromatosis generally presents as a painless swelling that has been present for less than a year, often fixed to the underlying muscle or bone but not to the skin (2, 4). The long axis of this tumor is usually oriented in the direction of the muscle bundle in which it is found; there is a tendency to infiltrate the surrounding structures, often encasing adjacent vessels and nerves without apparent invasion, possibly accounting for the clinical lack of pain (2, 3). Pathologically, these tumors lack a true capsule, although they may appear well circumscribed on imaging studies (6). Elongated, spindle-shaped cells of uniform appearance surrounded and separated by dense bands of collagen are noted (6). Mitoses are infrequent, with some variation in cellularity but no cells with atypical or hyperchromatic nuclei (6). The primary histologic differential diagnoses are those of a low-grade fibrosarcoma or reactive fibrosis (6).

Although aggressive fibromatosis is histologically benign, the biologic behavior of the lesion is similar to that of a low-grade malignant neoplasm, with local invasion and a high rate of recurrence. Recurrence is more common when the head and neck are involved (in as many as 70% of cases) than when a similar tumor is found in other locations; it is uncertain whether this is because of a more aggressive tumor or the technical difficulty of excision in this region (2).

Aggressive initial resection of aggressive fibromatosis with a wide margin of uninvolved tissue is necessary to prevent recurrence. Treatment, either before surgery or after postsurgical recurrence, has recently included the use of hormonal agents, as receptors have been demonstrated in these tumors (7, 8). Alternatively, both chemotherapy and radiation therapy have also been advocated (9, 10).

Although clinical reports of aggressive fibromatosis of the neck are available, there is a paucity of information concerning the imaging characteristics of this disorder. Two reports have been published demonstrating CT findings in similar sites involving the right carotid and right prevertebral spaces (2, 11). The lesions appeared hyperdense relative to surrounding muscle on contrast-enhanced scans in both cases (2, 11). However, no other CT data are available, and no MR signal characteristics in the neck have been described. Several studies have investigated the signal characteristics of intraabdominal and extraabdominal aggressive fibromatosis in other locations.

On CT examination, the masses can be poorly defined or well circumscribed and have variable attenuation before contrast enhancement (6). After contrast administration, the lesions are usually hyperdense relative to adjacent musculature, but can be isodense or hypodense (2, 6). On the CT examination in our case, the tumors also followed these more common attenuation characteristics and were slightly higher than adjacent muscle in attenuation.

The MR characteristics of aggressive fibromatosis of the extremities, mediastinum, and chest wall consist of poor margination, hypointensity or isointensity relative to adjacent muscle on T1-weighted images, and variable signal intensity on T2-weighted images (6, 12, 13, 14). Aggressive fibromatosis is most commonly predominantly intermediate in signal intensity, between muscle and fat on T2-weighted images, but occasionally may be lower in signal than muscle or higher than fat, as noted in our case (6, 12). Neurovascular and bone involvement is common (12).

Previous investigations also demonstrated areas of low signal on both T1- and T2-weighted images within the masses of aggressive fibromatosis that are thought to represent fibrous

tissue, areas of relative hypocellularity, or regions of increased collagen deposition within the tumor (12, 14, 15). The case described in this report demonstrated similar, irregular focal and linear areas of decreased signal intensity on all pulse sequences, suggesting fibrous tissue and agreeing with previously reported findings in other areas of the body.

Many of the imaging characteristics of aggressive fibromatosis are nonspecific, with the differential diagnosis including malignant tumors such as malignant fibrous histiocytoma, fibrosarcoma, and rhabdomyosarcoma; benign tumors such as neurofibroma and leiomyoma; and cervical fibrosclerosis (6, 16). Although heterogeneity of signal, poor margination, and neurovascular involvement are commonly seen in malignant tumors, the presence on all pulse sequences of internal areas of low signal intensity representing fibrous tissue or increased collagen deposition within the tumor should aid in differentiating aggressive fibromatosis from a malignant tumor, as this feature is uncommon in other soft-tissue tumors (12, 15). Although not seen in our patient, signal intensity lower than that of fat on T2-weighted images might also suggest that the diagnosis of aggressive fibromatosis, because most malignant soft-tissue tumors are higher in signal intensity (12).

In summary, although rare, aggressive fibromatosis should be considered in the differential diagnosis of any soft-tissue mass of the head and neck, particularly when the characteristic MR findings are present. In addition, once the diagnosis is suspected, the excellent anatomic detail provided by MR can aid in preoperative planning, because complete primary resection carries the highest likelihood of cure.

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